Health Care Provider Fact Sheet

Disease Name Sickle Cell Disease

Alternate Name(s) Sickle Cell Anemia

Acronym Hb S/S

Disease Classification Hemoglobinopathy

Symptom onset May be asymptomatic.

Symptoms Any sign of illness in an infant with sickling disease is a potential medical

emergency. Acute and chronic tissue injury can occur when sickled cells cause vascular occlusion. Sickling diseases can cause severe pain anywhere in the body, but most often in the hands, arms, chest, legs and feet. Complications may include, but are not limited to, the following: sepsis, acute chest syndrome, hand-and-foot syndrome, splenic sequestration crisis, aplastic crisis, stroke and painful episodes.

Natural history without treatment Infants with sickle cell disease are vulnerable to serious bacterial

infections that can be life threatening.

Natural history with treatment Reduced mortality and morbidity with penicillin prophylaxis.

Treatment The National Institutes of Health clinical guidelines for management of

sickle cell disease state, "Penicillin prophylaxis should begin by 2 months of age for infants with suspected sickle cell anemia, whether or not the definitive diagnosis has been established." Antibiotic therapy should

continue until at least 5 years of age.

Inheritance Autosomal recessive

General population incidence 1:500 for African American and Hispanic American.

OMIM Link http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=603903

Genetests Link www.geneclinics.org

Support Group Sickle Cell Information Center

http://www.scinfo.org/

Sickle Cell Disease Association of America, Inc.

http://www.sicklecelldisease.org

